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Gastroschisis in one twin neonate with extracorporeal liver

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ABSTRACT

We report a rare case of gastroschisis with extracorporeal liver suspected on late first trimester ultrasound and confirmed with second trimester ultrasound and magnetic resonance imaging in one fetus in a twin pregnancy. Liver herniation is common in omphalocele, a membrane-covered abdominal wall defect associated with other congenital anomalies. However, it is highly uncommon in gastroschisis, an uncovered abdominal wall defect aside of the cord insertion. Presence of liver herniation complicates prenatal differentiation between omphalocele and gastroschisis. The twins were born at 31 weeks' gestation due to preterm labor. The baby was treated with a silo device, followed by biologic mesh and a wound vac with instillation of fluid to prevent desiccation. Ultimately, the baby died of sepsis, with multiorgan failure and polymicrobial infection.

KEYWORDS Gastroschisis; giant gastroschisis; liver herniation; ruptured omphalocele

Gastroschisis is an abdominal wall defect usually found to the right of the fetal cord insertion without an overlying membrane. It is contrasted to the variable-sized midline defect of omphalocele, where the cord inserts on the membrane and organs are commonly extracorporeal. Occasionally, omphalocele membranes rupture, making it difficult to differentiate from gastroschisis. Extracorporeal liver involvement is rarely present in gastroschisis (2.3%–16%), leading to misdiagnosis as ruptured omphalocele.[1] We report another case of neonatal death secondary to gastroschisis with >50% extracorporeal liver herniation.

CASE DESCRIPTION

A 31-year-old woman presented for initial prenatal ultrasound at 12 weeks' gestation for a monochorionic, diamniotic twin pregnancy. Fetus B had an abdominal wall defect with extracorporeal liver serially evaluated by ultrasonography. At 20 weeks, the abdominal wall defect measured 1.4 cm to the right of the umbilical cord insertion. The stomach, bowel, and liver herniated through the defect *(Figure 1a)*. At 25 weeks, fetal magnetic resonance imaging confirmed a large abdominal wall defect with no membrane through which most of the small bowel, colon, stomach, gallbladder, and more than half of the liver herniated *(Figure 1b)*. At 31 weeks, a classical cesarean was utilized to allow adequate room to deliver the fetus and extracorporeal organs with careful attention to protect the fetal liver and its primary vasculature. To correct the defect, a 7.5 cm silo was modified and directly sewn to the fascia to slowly reduce the abdominal contents.

Due to poor progression with the silo, a biologic mesh was used as an onlay graft to cover the bowel and liver *(Figure 1c)*. A wound vacuum allowed the instillation of fluid to keep the mesh saturated, promote granulation tissue, and reduce the risk of intestinal fistula formation. The dressing was set to low-flow suction at 25 mm Hg. Dressing was changed every 3 to 5 days. The patient developed cellulitis, and cultures confirmed the offending organisms. Treatment required various combinations of antibiotics and switching irrigation to a sodium hypochlorite solution, which initially improved the infection. Despite these efforts, the patient developed renal failure with progressive edema, and multiple organ failure ensued. At 8 weeks of life, treatment concluded with withdrawal of care.

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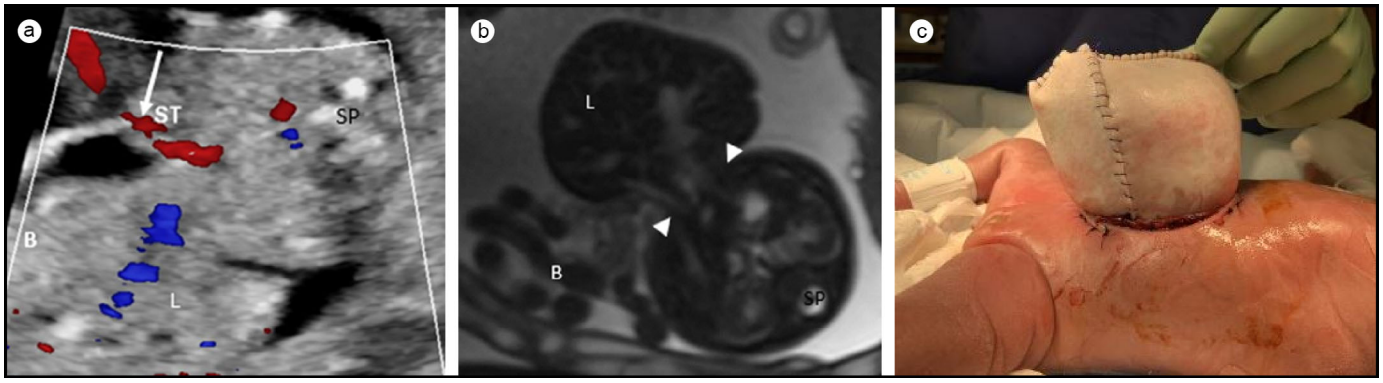

Figure 1. **(a)** Prenatal ultrasound imaging. Axial image at 20 weeks shows color flow in the umbilical cord to the left of the defect (arrow), as well as a portion of extracorporeal stomach (ST), bowel (B), and liver (L); the spine (SP) is labeled for orientation. **(b)** Fetal magnetic resonance imaging axial T2 HASTE at 25 weeks' gestation. Ventral abdominal wall defect (arrowheads) through which the majority of the liver (L) has herniated in addition to small and large bowel (B). The spine (SP) is labeled for orientation. **(c)** Two pieces of 8 × 16 cm Strattice mesh were trimmed to fit and circumferentially sewn to the abdominal wall.

DISCUSSION

The rarity of gastroschisis with liver involvement precludes large patient studies, so there is a need for case reports describing methods of treatment and outcomes. Gastroschisis has up to a 97% survival rate, but becomes nearly 100% fatal when associated with >50% of liver herniation.[1] The mortality rate for giant omphalocele is about 30% but can increase up to 66% based on other associated comorbidities such as pulmonary hypoplasia.[2] Rates for ruptured omphalocele are likely worse.[3] Location of the abdominal wall defect to the right of the umbilical cord insertion on prenatal imaging clinches the diagnosis.

Differentiation is important for prognosis and planning because omphalocele is associated with other congenital anomalies. In gastroschisis with extracorporeal liver and ruptured omphalocele, a large vertical midline abdominal incision and vertical uterine incision are prudent to allow for safe delivery to avoid damage to hepatic structures. The baby must be hemodynamically stabilized and the defect covered because the violated peritoneum raises risk of infection.

Eleven cases of gastroschisis with liver herniation have been reported, with five surviving with only minimal liver involvement.[1,4–6] This case supports the poor prognosis of babies with gastroschisis with extracorporeal liver, in this case with >50% of the liver out.

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